Rosset-Llobet, J., & Fàbregas-Molas, S. (2018). Rehabilitation and Plasticity of Task-Specific Focal Hand Dystonia. In D. Dressler, E. Altenmüller, & J. Krauss (Eds.), Treatment of Dystonia (pp. 256-260). Cambridge: Cambridge University Press. doi:10.1017/9781316459324.052



Chapter 51

# Rehabilitation and Plasticity of Task-Specific Focal Hand Dystonia

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## Historical Overview of Behavioural Methods

When we first encountered a musician with task-specific focal hand dystonia (TSFHD) in 1992, there was not much knowledge available about this disorder. At that time, many health professionals still believed this was a mental disturbance. Consequently, as there was no effective cure for this condition, most affected musicians had to end their professional careers.

Fortunately, in the 1990s, the scientific world became more aware of this field. Researchers demonstrated that the main cause of this disorder was not linked to any major mental problem (Sheehy & Marsden 1982). Because of this, TSFHD moved from psychology and psychiatry to neurology. To treat these patients, neurologists used pharmacological strategies including injections of botulinum toxin into the cramping muscles, as well as anticholinergic drugs. Although these treatments showed some temporary relief and improvement of symptoms, none of them provided a cure for the underlying causal changes (Altenmüller & Jabusch 2008).

Further research showed no brain damage in these patients; instead, patients exhibited organisational and functional changes of neuronal networks, including sensory processing, sensorimotor integration and motor planning on cortical and subcortical levels (Ridding et al. 1995; Bara-Jimenez et al. 1998; Elbert et al. 1998; Pujol et al. 2000; McKenzie et al. 2003; Quartarone et al. 2006; Tamura et al. 2009). It is quite probable that many of these alterations are interrelated. It could be that some alterations are the result of others, but do not cause TSFHD.

Because most of these central nervous system changes are considered to be the result of maladaptive changes to practice and since there is no structural brain damage, it seems logical that retraining, i.e. behavioural methods including instrumental practice and slow-motion exercises, should restore brain organisation and functionality. However, most patients report that any kind of basic relearning or intensive repetition of the affected movement to try to correct undesired responses (on the musical instrument if they have musician's dystonia, or doing calligraphy if they have writer's cramp) does not lead to consistent improvements, and can often exacerbate the problem.

Some researchers believe that focusing specifically on restoring some of the detected nervous system alterations could cure TSFHD symptoms. However, although they were able to improve or normalise some brain changes, and this was linked to clinical improvements, none of these protocols produced a complete, consistent and definitive resolution of the problem (Byl & McKenzie 2000; McKenzie et al. 2009; Rosenkranz et al. 2009). This could be due to several factors: the fact that they were retraining non-crucial changes in the brain; their focus on a symptom of TSFHD, not the cause; or the impact of the protocol not being strong and sustained enough to lead to a rearrangement of the disordered sensorimotor system.

At the same time, other researchers used more general and integrative procedures to treat TSFHD. This seemed to produce better results. In fact, for the first time, complete recovery in musicians having TSFHD was reported. Nevertheless, protocols were complex, required years of training, and the rate of success was low (Brockman et al. 1993; Tubiana & Chamagne 2000).

## Background of Our Retraining Approach

One of the most distinct features of TSFHD is the sensory trick. This phenomenon, which refers to sensory stimulation, e.g. wearing a latex glove while playing the instrument, is able to ameliorate or even completely restore motor control (Paulig et al. 2014). This kind of trick emphasises the important role of the sensory system as well as sensory motor integration in the pathophysiology of TSFHD. However, these kinds of tricks are not usually useful for retraining, as they have no lasting effects. Candia found a way around this limitation (Candia et al. 1999). He tested the efficacy of finger splinting as a new sensory input that should act as a more permanent sensory trick. He configured splints to place the fingers in a position different from the normal playing position. Candia also realised that splinting the compensatory fingers (those reacting to the original dystonic movement) is more effective than splinting the dystonic ones (for more information about how to identify dystonic and compensatory fingers, see Rosset-Llobet et al. 2013). Therefore, the first part of the daily retraining routine was to repeat different movement patterns on the musical instrument with the non-splinted fingers while some other fingers were splinted. Then, to avoid habituation effects, he used possibilities involving transfer. Transfer is the capability that allows the nervous system to use what has been learned in a specific context within another context, which has different conditions and task variants (for more details on transfer, see Seidler 2010). The second daily part of the retraining involved the repetition of the same

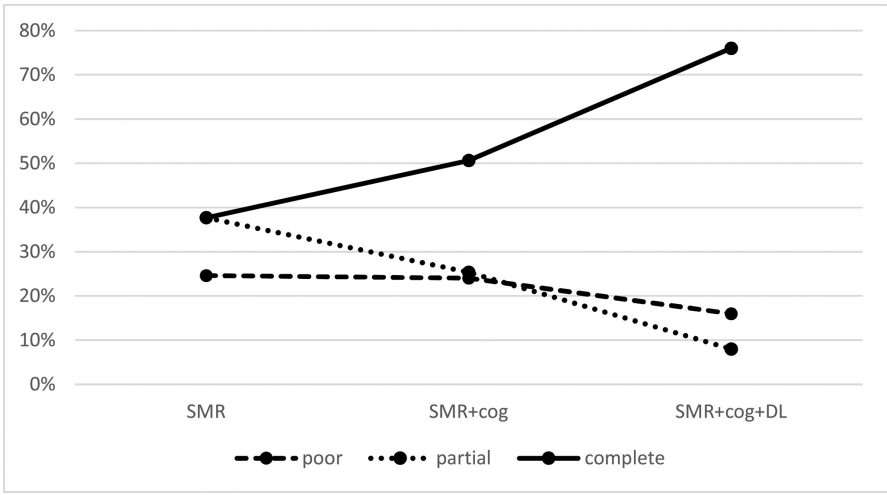


**Figure 51.1** Therapy results depending on the strategy used. We have grouped 230 consecutively treated patients into three groups depending on the protocol used: SMR alone, adding cognitive work to SMR, and combining these two strategies with differential learning (DL). Outcomes of patients were classified into three levels: poor, partial and complete recovery (see text for more details). Groups were compared using the *chi square* parametric test to analyse the significance. The confidence level used in all tests was 95 per cent. There were no statistically significant differences between group SMR and SMR + cog ($\chi^2 = 4.084$; $p = 0.130$) nor between SMR + cog and SMR + cog + DL ($\chi^2 = 5.339$; $p = 0.069$). There were statistically significant differences between SMR group and SMR + cog + DL ($\chi^2 = 13.302$; $p = 0.001$).

movement patterns, also on the musical instrument, but now without the splints, in order to transfer the new motor behaviour to the original (unaffected) situation. This procedure was called sensory motor retuning (SMR) and demonstrated a usefulness in reverting symptoms while causing parallel brain reorganisation (Candia et al. 2005). This was the technique that we adopted in 1999 to treat our TSFHD patients.

## Results from Our Training Without and With Additional Cognitive Therapy

We consecutively treated 130 musicians using this technique. To analyse the results, we have classified patients into three groups depending on their self-reported functional outcome. These are *complete recovery* (those who returned to instrumental activity with full recovery, with no dystonia symptoms at all), *partial recovery* (those with improvement of dystonia symptoms making a possible return to instrumental activity, but still feeling some functional limitation) and *poor result* (patients who dropped out of treatment or with an improvement not compatible with their musical career). Thirty-two of them (25 per cent) had *poor results*, 49 (38 per cent) *partial recovery*, and 49 (38 per cent) *complete recovery* (see Figure 51.1).

Although these figures constitute an improvement in comparison to other current rehabilitation techniques (Brockman reported 21 per cent successful results but without being clear how many of them had complete recovery), results are still far from acceptable (Brockman et al. 1993).

Patients with TSFHD frequently have certain psychological traits, including proneness to anxiety, perfectionism and obsessive compulsive tendencies. Some of these may be due to dystonia itself, but others probably existed before the onset of dystonia (Jabusch et al. 2004; Ioannou & Altenmüller 2014). We thought that this psychological status could account for the difference between the patients responding well to neurorehabilitation and those not responding to treatment. For this reason, our therapists receive training from a psychologist to be able to use cognitive techniques to improve these psychological factors that can interfere with therapy.

In a second study, we consecutively treated 75 musicians with TSFHD, adding cognitive behavioural therapy to SMR. From this group of patients 18 had *poor results* (24 per cent), 19 *partial recovery* (25 per cent), and 38 *complete recovery* (51 per cent). As can be seen in Figure 51.1, the proportions of patients changed, but the improvement was not statistically significant.

## Effects of tDCS on Retraining Results

Bearing in mind that patients with TSFHD exhibit greater activation of the primary sensorimotor cortex, deficits in surrounding inhibition and increased plasticity (Pujol et al. 2000; Hallett 2006), and knowing that transcranial direct current stimulation (tDCS) can safely modulate some of these aspects, we decided to explore the utility of combining this technique with SMR (Rosset-Llobet et al. 2014). As there are no studies demonstrating safety of tDCS if applied over longer periods, we decided to combine SMR and tDCS only during the first two retraining weeks. The results of this pilot study, subsequently confirmed by a larger survey, demonstrate that cathodal biparietal tDCS is a safe technique that enhances the improvement produced by neurorehabilitation on TSFHD and makes retraining easier (Rosset-Llobet et al. 2015). As the retraining protocol for these patients takes many months (average is 15 months), further studies are required to investigate whether two weeks of tDCS will improve not only short-term therapy results but also long-term results. It is also important to examine neurophysiological parameters in cases where brain stimulation is prolonged for more than two weeks. Due to lack of results from basic clinical research concerning the safety of long-term application of tDCS, we decided to stop using the treatment, although previous results have shown promise.

## Effects of Differential Training on Rehabilitation of Focal Dystonia

One intriguing characteristic of patients suffering from TSFHD is the stability of the dysfunctional behaviour after inception of dystonic symptoms. It is difficult to understand why, in a group

of patients with increased brain plasticity, retraining is so difficult. This question led us to explore approaches that have documented the ability to destabilise brain networks and promote learning. Differential training, also called differential learning (DL) and originally created to improve sports performance, is a technique which, by means of promoting and increasing system fluctuations, has also proven effective in rehabilitation (Schöllhorn et al. 2010). One of the strategies is to introduce noise to the task, constantly changing the way the task is done.

Recently, we have added this new approach to our protocol, introducing noise to SMR exercises. Patients were asked to change, every 3–5 seconds, the way they were performing the exercise, in order to interfere with the activity. For instance, they could change hand, arm or body position, modify the speed or intensity of a part of the movement, or perform it using other biomechanical patterns. We have completed treatment of 25 patients following this strategy (patients in progress are not included in this analysis). From this group, four had *poor results* (16 per cent), two *partial recovery* (8 per cent), and 19 *complete recovery* (76 per cent). These unpublished results are statistically better than those of the SMR group.

## Dynamic Systems Theory and Causes and Cures of TSFHD

From our point of view, the above-mentioned experiences open a new perspective to understand how TSFHD emerges and how it can be treated. First, there will not be a single technique useful for every patient, but a tailored combination of procedures, slightly different for each patient. We have seen that when we combine different strategies and adapt these strategies to each patient, results improve. However, we need to go a step further. TFSHD does not seem to have a unique patient profile; in other words, TSFHD patients do not exhibit the same psychological behaviour (Ioannou & Altenmüller 2014). Additionally, they do not exhibit the same behaviour in many other aspects, such as response to sensory tricks, their genetic predisposition, their biomechanical constraints, their gender, and, if they are women, their hormonal status (Rosset-Llobet et al. 2009, 2012; Lohmann et al. 2014; Paulig et al. 2014). In the near future we will need to more precisely identify these profiles and, accordingly, combine and adapt the different available techniques.

The second, and perhaps most challenging, consequence of this analysis is the necessity to see TSFHD from a completely different standpoint, not based on linear systems but rather on dynamic systems theory (Kurz & Stergiou 2004). According to this theory, the human body is an adaptable, complex and dynamic system capable of organising itself, although there is no one single factor inside the system capable of doing this task. Dynamic systems theory states that organisation of movement relies less on sensorimotor representations, and is also less hierarchical when considering how the multitude of interacting parts of the body are integrated into functional, goal-directed movements in dynamic environments. Research from this perspective has illustrated how spontaneous coordination tendencies provide neurobiological systems with the stability and the flexibility needed to adapt to complex, ever-changing environments (Kelso 1995). If a musician, due to his psychological behaviour or his training workload, shapes a very stable but rigid and inflexible nervous system, he or she will not easily be able to adapt to internal or external changes. This can lead to a degradation in the musician's performance or even to dysfunctional coordination when he or she faces challenging new situations or environments.

Interestingly, this way of seeing TSFHD can provide answers to questions that, until now, had no easy explanation. Musician's dystonia characteristically appears in high-level musicians that have practised a certain amount in their lives. Typically, dystonia emerges at a certain point in their careers, when they are under strenuous conditions (e.g. routine changes, physical problems, nerve conduction alterations, psychological stress). However, one of the urgent questions is: why do we occasionally find cases where only some, or even none, of these prerequisites can be identified (Rosset-Llobet et al. 2013)? From the dynamic system point of view, dystonia will appear when there is an imbalance between how able the musician is to adapt to inner or outer environmental changes (how flexible he or she is) and how intense these changes are. If he or she is flexible, the amount of environment changes necessary to produce system breakdown should be high. If he or she is very rigid, breakdown (dystonia, in this case) can occur more easily.

Even the role of genetics can be interpreted under this outlook. We know that 20 per cent of musician's dystonia patients have relatives with either musician's dystonia or writer's cramp (Schmidt et al. 2009). Genome-wide significance with musician's dystonia was observed for an intronic variant in the arylsulphatase G gene (Lohmann et al. 2014). This gene, along with others, could by means of molecular changes make the system more rigid and therefore more likely to suffer a breakdown when facing a destabilising environment. However, the data also suggest that genes are not the only factor causing the system to break under certain situations. TSFHD can be the result of a sum of many factors, genetics being only one of them. Other factors could compensate for the effects of genes, making the system more flexible. Or, conversely, the subject could never be subjected to a situation destabilising enough for the development of dystonia, even if the subject has a genetic predisposition.

Another poorly understood feature of musician's dystonia is the fact that women are less frequently affected (Lim & Altenmüller 2003). There is no evidence that this could be explained by a lower proportion of women playing musical instruments at a high level or by other occupational factors (Rosset-Llobet et al. 2005). We have demonstrated women having a normal menstrual cycle are less prone to suffer from musician's dystonia (Rosset-Llobet et al. 2012). We hypothesised in our paper that the physiological mechanism that protects them could be the hormonal cycle's capability to change cortical excitability and neuronal plasticity (Smith et al. 1999; Fernández et al. 2003), which could produce a protective effect. This hypothesis seems very plausible if viewed from the dynamic systems point of view. Cyclic female hormonal variations can facilitate brain adaptability to changes, promoting

system flexibility. When this is reduced (when a woman has a menstrual disorder) or lost (e.g. during menopause or pregnancy), women become more prone to TSFHD due to a reduction of the factors which make them more adaptable to their environment.

Finally, we need to explore in more depth the role that a differential learning and dynamic system approach can have in TSFHD neurorehabilitation and prevention. Both prevention and treatment can probably be done by means of rendering the nervous system more flexible and promoting network plasticity. This will make the nervous system more adaptable and, thus, less prone to dystonia, and also more able to restore its functional organisation. Here, we must develop tools to make rehabilitation possible and test these tools to confirm their impact on this group of patients.

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